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Abstract

The expression of emotions and internal states by individuals with severe or profound intellectual disabilities is a comparatively under-researched area. Comprehensive or standardised methods of assessing or understanding the emotions and internal states within this population, whose ability to communicate is significantly compromised, do not exist. The literature base will be discussed and compared to that within the general population. Methods of assessing broader internal states, notably depression, anxiety, and pain within severe or profound intellectual disabilities are also addressed. Finally, this review will examine methods of assessing internal states within genetic syndromes, including hunger, social anxiety and happiness within Prader-Willi, Fragile-X and Angelman syndrome. This will then allow for the identification of robust methodologies used in assessing the expression of these internal states, some of which may be useful when considering how to assess emotions within individuals with intellectual disabilities.

Keywords: intellectual disabilities, emotion, internal state, learning disabilities, behavioral phenotypes, hunger, pain

The Expression and Assessment of Emotions and Internal States in Individuals with Severe or Profound Intellectual Disabilities

Introduction

The focus of this review is the expression of emotion and internal states by individuals with severe or profound intellectual disabilities and the corresponding methods of assessment. A review of this area is important in order to evaluate the current state of empirical literature and highlight the need for further research. It will be argued that researchers and clinicians should be proficient at directly assessing and interpreting the expression of internal states of individuals within this population whose ability to communicate is, by definition, significantly compromised. Expertise in this area would reduce reliance on informant opinion when making decisions with regard to diverse and fundamental issues such as physical health, choice and quality of life (see Ross & Oliver, 2003).

This paper opens with a broad overview of the expression of internal states and emotions; their development and role within infancy. The review will then focus on expression of emotions within individuals with severe or profound intellectual disabilities. A systematic review is then described with critique of methodology. Physical and behavioral (e.g. body posture) indicators of internal states will be discussed and evaluated and the clinical implications considered. Due to the paucity of the literature on emotions, the review will then be extended to consider methods of assessing internal states such as anxiety, depression and pain within this population.

Finally, the review will highlight the range of methodologies and measures used to assess internal states within individuals with genetic disorders[1] associated with intellectual disabilities. This is a valuable and rich literature base as many genetic disorders have well-documented atypical internal states, including excessive hunger and social anxiety, that have been successfully researched using a variety of methods. Each of these methods could further inform the literature regarding emotions and emotional expressions within individuals with severe or profound intellectual disabilities.

Whilst there is a substantial body of literature discussing the capacity of individuals with intellectual disabilities to recognise or label other people's internal states, this will not be covered within this paper. For reviews see Owen, Browning and Jones (2001) and Zaja and Rojahn (2008).

1.1 Basic internal states and emotions

There has been debate within the literature as to what constitutes the "basic" or "fundamental" human emotions. The widely accepted basic human emotions are happiness, fear, anger and sadness, with disgust and surprise also accepted by most (Turner, 2000) but there is little consensus concerning interest, anticipation, guilt and shame. Using mostly anecdotal evidence, Darwin (1872) suggested that since the expression of basic emotions, particularly facial expressions, are similar in humans around the world, they must have a hereditary basis. He proposed that emotions serve a function and therefore should be seen as adaptive and, arguably, essential to the survival and reproduction of species. Turner (2000) has expanded upon this work, suggesting that behavioral and facial expressions are critical to group harmony as they effectively communicate emotions, feelings and intent. This emotion-

[1] For the purpose of this review, the term genetic disorders refers to intellectual disabilities with an identifiable genetic aetiology (as oppose to idiopathic intellectual disabilities)

signalling hypothesis is attracting growing interest and support (e.g. Consedine, Magai, & Bonanno, 2002).

1.2 Facial expressions of emotions

Facial expressions of emotions play a key role in early development as they allow for communication between the pre-verbal infant and their caregiver (Spangler, Emlinger, Meinhardt, & Hamm, 2001). In order to quantify and use the information afforded by facial expressions shown by infants for the purpose of research, reliable, valid and efficient coding systems must be used (Cohn, Zlochower, Lien, & Kanade, 1999). Two systems, both of which require extensive training procedures, code observable changes in the face: the System for Identifying Affect Expression (AFFEX; Izard, Dougherty, & Hembree, 1983; formerly known as the Maximally Descriptive Facial Movement Coding System (MAX; Izard 1979) and the Facial Expression Scoring Manual (FESM; Izard, 1971; 1977)) and the Baby Facial Action Coding System (Baby—FACS; Oster, 2000; based upon the Facial Action Coding System (FACS; Ekman & Friesen, 1978)). Using various methods, good face and current validity are reported (Ekman, Friesen & Ancoli, 1980).

Using these coding systems, researchers have begun to identify the development of facial expressions in neonates and babies. Although crying is usually the first demonstration of an emotion (Oster, Hegley, & Nagel, 1992), the associated facial expression is usually coded as undifferentiated distress (Oster *et al.*, 1992). The earliest facial expressions of emotions seen in newborns (aged 3-10 hours) are in relation to taste; both positive in response to sucrose and negative in response to quinine solutions (Steiner, Glaser, Hawilo, & Berridge, 2001).

Increasing complexity of emotion expression emerges throughout the first two years of life. Using the FESM, basic emotions can be elicited in infants ranging in age from one to

ten months (Hiatt, Campos, & Emde, 1979; Izard, Huebner, Risser, McGinnes, & Dougherty, 1980). These emotions and expressions become functionally organised in relation to the infant and their social context (e.g. Bennett, Bendersky, & Lewis, 2005), resulting in responses that are more differentiated and specific (e.g. Witherington, Campos, & Hertenstein, 2001). This coincides with the development of several important cognitive concepts, including understanding others' intentions (Izard, Hembree, Dougherty, & Spizzirri, 1983), a concept of self, emotion regulation (Stifter & Spinrad, 2002) and elaboration of an internal "affective map" (e.g. Gendler-Martin, Witherington, & Edwards, 2008). Infants begin to label their own emotions from 18-20 months of age (Bretherton, Fritz, Zahn-Waxler, & Ridgeway, 1986), a skill that is highly correlated with cognitive abilities (Bennett *et al.*, 2005) and social behavior (e.g. Fine, Izard, Mostow, Trentacosta, & Ackerman, 2003; Izard *et al.*, 2001).

Alongside having a strong communicative function, facial expressions influence the perceived attractiveness of an infant (Power, Hildebrandt, & Fitzgerald, 1982) and consequently affect adults' reactions to them (Hildebrandt & Fitzgerald, 1978). For example, children rated as more physically attractive are treated more favourably (e.g. Adams, 1977). Infants showing positive facial expressions are also rated as "cuter" (Hildebrandt, 1983), although it is unknown if or how flexible perceptions of cuteness, and therefore adults' behaviors, are in response to changes in facial expressions.

1.3 Populations within which facial expressions of emotions are altered or impaired

Despite the strong similarities of facial expressions of emotions between and within cultures and even species, some populations show impairments. These include individuals with schizophrenia, dementia, Möbius syndrome and those with severe or profound

intellectual disabilities. Literature pertaining to the expression of emotions within each of these populations will be reviewed briefly below.

1.3.1 Individuals with schizophrenia. Deficits in the expression of emotions are well documented within schizophrenia (e.g. Kring & Neale, 1996; Salem, Kring, & Kerr, 1996; Tremeau et *al.*, 2005) using a range of methodologies (e.g. self-ratings; Schneider, Gur, Gur, & Shtasel, 1995; informant rating scales; Andreasen, 1984a,b; facial coding systems; Berenbaum & Oltmanns, 1992). The significant impairments in facial expression in individuals with schizophrenia are not due to reduced emotional experience; individuals report emotional experiences equal to or possibly greater than individuals without a psychiatric diagnosis (Herbener, Song, Khine, & Sweeney, 2008; Kring, Kerr, Smith, & Neale, 1993), nor are they simply due to side effects of medication (Earnst *et al.*, 1996; Putnam & Kring, 2007). Despite little outwardly observable facial expression, sensitive methods (e.g. electromyography) have identified greater activity in muscles responsible for smiling and frowning in response to positive and negative stimuli respectively (Earnst *et al.*, 1996). There are also recognised deficits in posed emotional expressions; although varying methodologies makes it difficult to draw firm conclusions (see Izard & Dougherty, 1982, for a discussion).

Such difficulties in both spontaneous and posed expressions alongside the dissociation between experience and expression of emotions suggests an underlying physical or neuromotor deficit (Dworkin, Clark, Amador, & Gorman, 1996) rather than a specific deficit in neural or behavioral systems that underpin the experience (Putnam & Kring, 2007). Despite this hypothesis, there has been little research published on effective interventions, medical or psychological, within this area.

1.3.2 Individuals with dementia. Facial expressions of emotions are important communicative tools (Jakobs, Manstead, & Fischer, 1999), particularly in those who are pre-verbal (see Nelson & de Haan, 1997) or for those with impaired speech (Hemsley *et al.*, 2001), such as individuals who develop dementia. As dementia, most notably Alzheimer's disease, progresses, declines are noted in expressive language skills (Faber-Langendoen *et al.*, 1988; Kertesz & Clydesdale, 1994) and consequently the ability to report internal affective states (e.g. pain; see Smith, 2005, for a comprehensive review). Individuals must therefore rely on other methods to communicate.

Facial expressions, both "adaptive" and "inappropriate" (Porter *et al.*, 1996) are enhanced in frequency and intensity in individuals in the early stages of dementia when compared to healthy controls (e.g. Kunz, Scharmann, Hemmeter, Schepelmann, & Lautenbacher, 2007). As the dementia advances, facial expressions gradually decrease (Norberg, Melin, & Asplund, 1986); by end stage dementia it is only possible to identify individual fragments of facial expressions using the FACS. However, even at this late stage, pleasant and unpleasant stimuli evoke differences in heart rate, respiratory rate and skin temperature (Asplund, Norberg, Adolfsson, & Waxman, 1991).

Akin to individuals with schizophrenia, it could be concluded that individuals with end stage dementia may still experience various emotions, but have difficulties expressing them due to neurological change. The task therefore for researchers and clinicians is to develop methods of reliably assessing these states within these populations.

1.3.3 Individuals with Möbius syndrome. Möbius syndrome is a rare congenital disorder associated with unilateral or bilateral paralysis of the abducen and facial cranial nerves (Möbius, 1888, cited in Gillberg & Steffenburg, 1989), although involvement from other cranial nerves is common. It is sometimes associated with a mild degree of intellectual disability and co-morbid autism (e.g. Gillberg & Steffenburg, 1989). This facial paralysis, evident from early infancy, means that individuals with this syndrome cannot produce facial expressions of emotions and often have delays or difficulties with speech (Meyerson & Foushee, 1978).

Surprisingly, there are few studies investigating methods of emotional expression within individuals with Möbius syndrome. Sjögreen, Andersson-Norinder, and Jacobsson (2001) observed impairments in facial expressions in all 25 of their participants (aged 2 months to 55 years), ranging from a mild isolated unilateral weakness to profound bilateral paralysis. Szajnberg (1994) visited one child at six, ten and fourteen months of age and observed interactive play with both parents. He suggests that whilst the child was developing and adapting alternative ways of affective communication (such as hand gestures and body posture), the lack of facial expressions began to effect social interactions when the child wanted to express more complex feelings. In line with theories of emotional development, impairments in recognising others' facial expressions are also noted (Calder, Keane, Cole, Campbell, & Young, 2000; Giannini, Tamulonis, Giannini, Loiselle, & Spirtos, 1984).

Cole (2008) discusses the potential impact that difficulties with expression may have on the experience and development of complex emotions. Surgery and biofeedback mechanisms are continually being trialled and reported but with small samples only (e.g. Zuker, Goldberg, & Manktelow, 2000). These areas require further investigation, combining the results of multiple methodologies (e.g. scanning, self-report, observations) in order to

reach valid conclusions which may help inform interventions within this, and other populations, for whom emotional expressions are impaired.

1.3.4 Individuals with severe or profound intellectual disabilities. Understanding and assessing emotions of individuals with intellectual disabilities[2] is within its infancy (Clark, Reed, & Sturmey, 1991). There is considerable debate within the literature as to whether individuals with intellectual disabilities express the same range of emotions in the same way as people without intellectual disabilities. This debate has in part arisen from differences based on level of intellectual disability and, importantly, differences based upon the aetiology of the intellectual disability. What has not been ascertained is whether there is a difference in the experience or expression of emotions within individuals with intellectual disabilities.

We now review the literature base pertaining to the expression of emotions within individuals with severe or profound intellectual disabilities. The quantitative results of a comprehensive literature search are presented and compared to searches undertaken within sub-populations of the general population. The literature is then summarised, describing facial and behavioral expressions of emotions within this population alongside the ability of others to recognise and label such expressions. The expression of emotion within individuals on the autistic spectrum is not discussed specifically within this review but is reviewed by Bormann-Kischkel, Amorosa, and von Benda (1993).

[2] The term "intellectual disability" will be used throughout this thesis in preference to a number of other terms (learning disability, mental retardation, mental handicap and mental deficiency) in line with current usage of terminology in recent Department of Health publications

2 Literature Search

2.1 Method

Computerised searches were undertaken on Web of Knowledge and Medline. Web of Knowledge searches the titles and abstracts of all journals listed within the Science Citation Index, Social Sciences Citation Index and the Arts and Humanities Citation Index published after 1981. MEDLINE searches the abstracts and titles of over 1,800 journals published by Elsevier.

The specified keywords were learning disabilit* (spelt to retrieve articles with the singular or plural spelling), intellectual disabilit*, developmental disabilit*, mental retardation, mental handicap and mental deficiency. These were combined with emotion*, affect, mood, facial express*, happy, sad, disgust, surprise, anger and fear. Abstracts were selected if they provided information on expressions of internal states in individuals with severe or profound intellectual disabilities. Once this search was completed, reference lists of the selected papers were also searched. The results of the searches and information retrieved are summarised below.

2.2 Results

The electronic searches on Medline and Web of Knowledge yielded 2544 papers. However, only sixteen of these focussed on expression of emotions in individuals with severe or profound intellectual disabilities. Dates of publication ranged from 1987 to 2008. Eight studies were conducted in the UK, six in the USA, one in Hong Kong and one in New Zealand. Methodologies employed included behavioral and facial observations, informant based interviews and questionnaires.

The main topics covered by the 2528 papers that did not focus on the expression of emotions in individuals with severe or profound intellectual disabilities were: the ability of individuals with intellectual disabilities to recognise others' emotions, the emotions of family members or individuals working with people with intellectual disabilities, the expression of emotions in individuals with mild or moderate intellectual disabilities and the adaptation of therapeutic approaches (e.g. cognitive behavioral therapy) for working with individuals with mild or moderate intellectual disabilities.

2.3 Comparison with typically developing population

In order to evaluate the volume of literature pertaining to the expression of emotions within individuals with severe or profound intellectual disabilities, comparison searches were undertaken for two populations. Firstly, neonates (i.e. under one month old) were chosen as they are a population with a discrete body of literature in which the problem of lack of verbal communication is shared. Statistically, they also make up approximately the same proportion of the population (.128% based on data from the 2001 UK census) as individuals with intellectual disabilities (.13%). For this search, the specified keywords of neonate and newborn were combined with the emotion related keywords described above (emotion, affect, mood, facial expression*, happy, sad, disgust, surprise, anger and fear).

The second population used for comparison was infants; i.e. under 12 months. This population, although larger in proportion (approximately 1.5% based on data from the 2001 UK census), is similar to individuals with severe or profound intellectual disabilities in that they rely on non-verbal methods to express their emotions. Here, the keyword of infant was combined with the emotion related keywords listed above.

The electronic searches on Medline and Web of Knowledge yielded 12350 papers for neonate and emotional keywords, and 14679 papers for infant and emotional keywords. For

reasons of practicality, further searches were not undertaken to identify what percentage of these papers specifically addressed the expression of emotions. However, the number of papers identified suggests that the literature regarding the expression of emotions in individuals with intellectual disabilities and, more specifically, severe or profound intellectual disabilities, is comparatively sparse.

3 Expression of emotions and internal states in individuals with severe or profound intellectual disabilities

3.1 Defining severe and profound intellectual disabilities

The DSM-IV (American Psychological Association, 1994) and the ICD-10 (World Health Organisation, 1992) classify intellectual disability into four categories based upon IQ (or estimates thereof): mild (IQ range 50-70), moderate (IQ range 35-49), severe (IQ range 20-34) and profound (IQ below 20). This review will focus upon literature and research with individuals with severe or profound intellectual disabilities as both language and sensory deficits (the importance of which will become apparent throughout this review) are predominant within this population.

3.2 Methods of identifying emotions within individuals with severe or profound intellectual disabilities

There is now an extensive literature describing the cognitive functioning, adaptive behavior, challenging behavior and service planning needs of individuals with severe and profound intellectual disabilities. The question must be posed as to why research has not addressed the emotions experienced and expressed within this population, given their perceived importance in other populations (Arthur, 2003; Bailey, Matthews, & Leckie, 1986; Wagner, 1991).

The papers identified in the above literature search (alongside other supporting papers) will now be reviewed. This will allow an overview of the current literature base whilst highlighting some of the methodological difficulties that may have limited progress within this area.

3.2.1. Facial Expression of Emotions. Facial expressions are powerful nonverbal channels for conveying emotions (Plesa-Skwerer, Faja, Schofield, Verbalis, & Tager-Flusberg, 2006). Reviewed in section 1.2, facial expressions of emotions are expressed by typically developing infants at a very early age (Steiner *et al.*, 2001) and can be reliably interpreted by both trained and untrained adults who are familiar or unfamiliar with the infant (Hiatt *et al.*, 1979; Sorce & Emde, 1982).

Deficits in expression of emotions are noted within individuals with idiopathic and genetically determined intellectual disabilities (e.g. Down syndrome; Cicchetti & Sroufe, 1976; Cornelia de Lange syndrome; Johnson, Ekman, Friesen, Nyhanm, & Shear, 1976; autism and idiopathic intellectual disabilities; Yirmiya, Kasari, Sigman, & Mundy, 1989). There are two important themes to consider when reviewing this literature: the capacity of individuals with severe or profound intellectual disabilities to communicate emotions through facial expressions; and, perhaps more importantly, the ability of individuals working or living with that individual to interpret and understand such expressions.

3.2.1.1. Facial expressions in individuals with severe or profound intellectual disabilities. There is very little literature investigating the development of expressions of emotions within this population with regard to chronological or mental age. Cicchetti and Sroufe (1976) found a positive association between cognitive and affective development in a longitudinal sample of fourteen infants with Down syndrome. The developmental trajectory of laughing to increasingly complex stimuli was the same as a

sample of typically developing infants, but was delayed by a number of months dependent upon degree of intellectual disability. Interestingly, they noticed that those with a greater degree of intellectual disability may only smile under conditions where the more able infants would laugh, suggesting that the expression of emotions may be somewhat more subtle in children within this population. Cicchetti and Sroufe suggest a motor co-ordination element to this delay; research into more able children with Down syndrome has identified specific deficits in eliciting a reliable facial expression of fear (see Sroufe & Wunsch, 1972), although this has not been investigated within individuals with severe or profound intellectual disabilities.

Reliable and valid methods for observing facial expressions may allow clinicians, researchers and caregivers to develop a better understanding of how an individual with very limited or no expressive language might convey a positive or negative affective state. This could then reduce the reliance on informants when making important or complex decisions (Ross & Oliver, 2003) as even using structured questionnaire measures, informants' reports differ from those of individuals with intellectual disabilities, especially in relation to more complex internal experiences such as those associated with mental health (Ruddick & Oliver, 2005).

Within the behavioral literature, there are few studies that have observed facial expressions to obtain information about the individual's experience of the intervention (see Ross & Oliver, 2003, for a full review of these studies). Lindauer, DeLeon, and Fisher (1999) observed facial indices of positive and negative affect when evaluating the effects of an enriched environment programme. Oliver, Hall, Hales, Murphy, and Watts (1998) noted changes in positive and negative affect before and after an intervention to reduce self-

injurious behavior. Facial expressions of emotions have also been used to identify reinforcers or punishers (e.g. Fisher *et al.* 1992).

Green and Reid (1996) operationally defined facial expressions of happiness and unhappiness in order to evaluate a program aimed at increasing happiness in individuals with profound intellectual disabilities. Whilst happiness is a private event that is not easy to measure directly or manipulate (Kennedy & Souza, 1995), Green and Reid suggest that an individual's public behaviors can give them "clues" as to their private experience. This correlational approach does not allow for definitive conclusions as to whether happiness itself was altered and observed (Green & Reid, 1996) but does provide a starting point for observing such expressions of emotions within this population. Green and Reid increased facial indicators of happiness through their "fun time" program; results which have since been replicated by Davis, Young, Cherry, Dahman, and Rehfeldt (2004) and Ivancic, Barrett, Simonow, and Kimberly (1997).

This work by Green and Reid (1996; 1999a,b) would benefit from further replication, as at present it is limited by small participant numbers and thus potential lack of generalisability. Replicating the interventions with individuals with mild or moderate intellectual disabilities, using self-report or questionnaire methods alongside observations or behavioral expressions of choice, could also provide validation for using such behavioral indicators of happiness.

3.2.1.2. Recognition of facial expressions in individuals with severe or profound intellectual disabilities. The ability to provide reliable, independent indices of emotions becomes pertinent when the literature regarding caregiver recognition and interpretation of facial expressions of emotions in individuals with severe or profound

intellectual disabilities is reviewed. Again, a paucity of research within this area limits the extent to which conclusions can be drawn.

The facial expressions of children with severe intellectual disabilities in static photographs are less well recognised than those of typically developing children (Maurer & Newbrough, 1987a), with happy faces being easier to recognise than neutral, sad and angry faces. Adults inexperienced with individuals with intellectual disabilities were less accurate on all four emotions than parents of children with intellectual disabilities as they tended to label most facial expressions as "neutral" (Maurer & Newbrough, 1987b). Maurer and Newbrough suggest that parents may be used to interpreting more subtle expressions of emotions that other adults may overlook. Interestingly, there was no difference between parents and inexperienced adults on rating emotions of typically developing children, suggesting that the differences found were not due to general emotion recognition abilities. The authors conclude that children with intellectual disabilities do express their emotions through facial expressions, but experience and careful observation is required in order to interpret and understand them.

It could be argued that interventions within this field should be aimed at improving emotion recognition within families and caregivers as they are best placed to learn and implement new skills (Butterfield & Arthur, 1995). Such caregiver based interventions have previously proven successful (e.g. Singh *et al.*, 2004) but are hampered by high levels of staff turnover and burnout (Hastings, Home, & Mitchell, 2004); limiting the extent to which such interventions are practical or beneficial.

3.2.2 Behavioral expression of emotions. Developing an understanding of how behavior relates to affect or emotion is important in individuals with few or no verbal skills (Helm, 2000; Ross & Oliver, 2003). Challenging behavior has been associated with communication difficulties within both experimental (e.g. Bott, Farmer, & Rhode, 1997; Quine, 1986) and carer or support staff based questionnaire studies (e.g. Hastings, 1997). This has led to the suggestion that challenging behavior may be analogous to communicating expressions of emotion or emotional distress (e.g. Carr & Durand, 1985; Thurman, 1997).

Lindauer *et al.* (1999) found a high correlation between indices of negative affect and challenging behavior in their case study of a 23-year-old woman with severe intellectual disabilities and depression. She engaged in self-injurious behavior in 36% of the ten-second intervals in which she displayed negative affect and only 3.2% of the intervals where she did not display negative affect. However, the authors did not conduct a lag analysis in order to establish the sequence of and relationship between these behaviors. Carr, McLaughin, Giacobbe-Grieco, and Smith (2003) also found a higher likelihood of problem behavior during an intervention if staff rated a participant as being in a "bad mood" earlier in the day. Interestingly, they also note that offering the participant access to stimuli associated with positive moods during the intervention improved mood ratings.

The literature does seem to suggest a relationship between emotions and challenging behavior, although, at present, it is unknown if this is causative or due to an underlying mediating factor, such as physical pain (see an excellent review by Carr & Owen-DeSchryver, 2007).

3.3 Methods of assessing internal states in individuals with severe or profound intellectual disabilities

This review has so far established that individuals with severe and profound intellectual disabilities experience a range of emotions and express them through facial expressions and behavior. Such conclusions are mainly based upon observational studies (e.g. Green & Reid, 1996) or independent ratings of videos or photographs (e.g. Maurer & Newbrough, 1987a,b). Due to the paucity of research focussing explicitly on the expression of emotions within individuals with severe or profound intellectual disabilities, the remit of the review will be broadened to include other "internal states" including mood, hunger and affect. Including these broader internal experiences will allow for the discussion of methods of assessment that could potentially be used or adapted to assess emotions within this population.

Due to the degree of intellectual disability, methodologies widely used within the typically developing population are often inappropriate for assessing internal states with individuals with intellectual disabilities. The Interact Short Form – People with Profound Intellectual Disabilities Edition (original version by Baker & Dowling, 1995; adapted by Liu *et al.*, 2003) is an eleven-item observation tool designed to assess internal states before and after interventions or environmental changes. A trained rater observes the individual while performing daily routine sedentary activities (Baker & Dowling, 1995). Internal consistency (.83) and inter-rater reliability (mean Kappa=.72, range .68-.77) are both good although they were based upon a small sample (n=33 for inter-rater reliability) with very limited variation in mood and behavior.

Comparatively more assessment methods are available for assessing broader internal states, notably depression, anxiety, and pain. Each of these areas will be briefly reviewed below in

terms of their presentation (in relation to the typically developing population) and current methods of assessment. A full review of all available assessments within these areas is outside the scope of this review; comprehensive reviews are provided elsewhere for mood (Ross & Oliver, 2003), anxiety (Matson, Smiroldo, Hamilton, & Bagilo, 1997a) and pain (Symons, Shinde, & Gilles, 2008).

3.3.1 Depression and anxiety. It was only recently accepted that individuals with intellectual disabilities are more susceptible to mental health problems than the typically developing population (e.g. Matson, Gardner, Coe, & Sovner, 1991). At present, estimate rates for mental health problems range from 10 to 60% for individuals with intellectual disabilities (King, State, Shah, Davanzo, & Dykens, 1997). Such high levels of variability are reported due to sampling biases, methodological or diagnostic differences and psychosocial and environmental factors (see Ross & Oliver, 2003). Prevalence rates within children with severe or profound intellectual disabilities are yet to be determined.

Whether depression and anxiety present differently among individuals with severe and profound intellectual disabilities remains a matter of debate. Due to the communication difficulties inherent within this population, obtaining subjective reports of feelings, fears and thoughts is compromised or impossible (Matson *et al.*, 1997a). Reliance is therefore placed upon rating scales, informant interviews and potentially, observational techniques (each of which is described in detail elsewhere (e.g. Bramston & Fogarty, 2000; Liu *et al.*, 2007; Ross & Oliver, 2003)) to identify changes in sleep (e.g. Marston, Perry, & Roy, 1997), appetite and low mood (e.g. Charlot, Doucette, & Mezzacappa, 1993).

Matson *et al.* (1997b) highlight some of the problems with differential diagnosis when assessing mental health problems within individuals with severe or profound intellectual disabilities. They highlight numerous symptoms highly suggestive of anxiety within the

general population that occur frequently within individuals with severe or profound intellectual disabilities without anxiety. These include sudden motor or vocal responses (which may be part of a stereotypic behavior), trembling or shaking (which may be associated with tardive dyskinesia associated with psychotropic medications; Aman & Singh, 1985; 1991), sleep difficulties and exaggerated startle responses (present in many individuals with autism; Schopler, Reichler, & Renner, 1988). Diagnoses should therefore always consider information from informant-based questionnaires alongside a detailed history of behavior and behavioral change.

3.3.2 Pain. Pain is one of the most common experiences of all humans and is, arguably, one of the most difficult to identify (Phan, Edwards, & Robinson, 2005). Akin to the other internal states and emotions, it is a subjective, internal experience that cannot be assessed through direct measurement. The neuroanatomical system required to feel pain is complete by 26 weeks gestation, however, the developmental processes necessary for the mindful experience of pain are yet to occur (see Derbyshire, 2006). This, along with the facial and behavioral expression of pain, develops throughout childhood, and is shaped by cultural norms (e.g. Schiavenato *et al.*, 2008).

The expression of pain among individuals with severe or profound intellectual disabilities has received considerable levels of attention. Due to degree of intellectual disability, difficulties are present in self-reporting the presence of pain, its intensity (Biersdorff, 1994) and location (Hennequin, Morin, & Feine, 2000). This, coupled with a common misconception that all individuals with intellectual disabilities are insensitive or indifferent to pain (see Foley & McCutcheon, 2004, for a review), highlights the need for sensitive assessment methods and integrative translational research within this area (Symons *et al.*, 2008).

There are many instruments that translate the internal experience of pain into quantitative data, including some that have been developed for individuals with severe or profound intellectual disabilities. Defrin, Pick, Peretz, and Carmeli (2004) suggest that behavioral indices of pain can provide a more complete picture of the pain experience in individuals with severe or profound intellectual disabilities. Vocalisations, verbalisations, physical gestures, autonomic responses, general appearance and facial expressions can provide information on the degree of pain and its associated discomfort. Self-injurious behavior is also associated with higher scores on pain measures (Symons, Harper, McGrath, Breau, & Bodfish, 2009).

Bodfish, Harper, Deacon, and Symons (2001) developed the Pain and Discomfort Scale (PADS) to assess pain in individuals with little or no expressive language. The scale relies upon facial expressions and body movements to indicate acute pain and discomfort. Validation studies show promising results for both acute and chronic pain (Bodfish *et al.*, 2001). Phan *et al.* (2005) conclude that although the instrument is somewhat difficult to use, it is better than using clinical observations alone. Davies and Evans (2001) conclude that the best method of assessing pain in individuals with severe or profound intellectual disabilities is a thorough clinical assessment combined with a familiarity and understanding of both intellectual disabilities and the individual themselves. As pain is likely to be expressed through more than one non-verbal behavior (Davies & Evans, 2001), and may differ between chronic and acute pain (Collignon & Giusiano, 2001), it is important that caregivers and clinicians alike remain flexible with their interpretation and approach.

3.3.3 Summary. The studies reviewed above suggest that individuals with severe and profound intellectual disabilities do experience some extremes of internal states, such as depression, anxiety and pain, but the presentation of these experiences may differ slightly due to the degree of disability. The underlying theme from these three areas (depression, anxiety, and pain) is the importance of recognising and acknowledging facial and behavioral markers of internal states within this population. Structured behavioral observation schedules with acceptable validity and reliability are required in order to utilise the communicative nature of such expressions of internal state and experience.

3.4 Assessment of internal states within individuals with intellectual disabilities of a genetic aetiology

This review has so far identified a lack of literature relating to the expression of emotions in individuals with severe or profound intellectual disabilities. Methods of assessing broader internal states, including depression, anxiety and pain are somewhat better, but remain in their infancy.

If, as is widely accepted within the literature, internal states and emotions have a genetic element (albeit one that interacts or can be sometimes overridden by the environment), it could be hypothesised that differences in internal states, or their expression, may be present in individuals with genetic, or chromosomal, abnormalities, such as those with intellectual disabilities with genetic aetiologies.

Genetic aetiologies can be identified for approximately 13-46% of those with a mild intellectual disability and 33.2-70% of individuals with severe or profound intellectual disabilities (see Leonard & Wen, 2002, for an in depth discussion). Many individuals with genetic aetiologies will have a profile of behavioral (and physical) features that are highly

associated with their genetic cause of intellectual disabilities. Such associations have led to the notion of the "behavioral phenotype", which proposes that the genetic basis of specific intellectual disabilities (syndromes) strongly influences the comparative likelihood of the occurrence of particular behaviors (Dykens, 1995).

The literature pertaining to behavioral phenotypes within individuals with genetic disorders and intellectual disabilities contains descriptions of behaviors that have been researched in depth using multiple methods, which ultimately combine to derive multi-level models. By turning to this literature within this review, we can identify specific behaviors and internal states that have been researched in depth within syndrome-specific populations. This then allows us to identify robust methodologies used in assessing the expression of these internal states, some of which may be useful when considering how to assess emotions within individuals with intellectual disabilities. Three examples of this are discussed in turn; hunger in Prader-Willi syndrome, social anxiety in Fragile-X syndrome and laughing and smiling in Angelman syndrome.

3.4.1 Assessment of hunger in individuals with Prader-Willi syndrome. Prader-Willi syndrome is a genetic syndrome that arises from the absence of the expression of the paternal genes on chromosome 15q11-q13 (see Boer *et al.*, 2002). Current estimates suggest a prevalence of approximately 1 in 25,000 births and a gender ratio of 1:1 (Whittington *et al.*, 2001). The behavioral and cognitive phenotype has recently been reviewed (see Dimitropoulos *et al.*, 2000) and includes low-average intelligence or a mild to moderate intellectual disability, excessive eating, repetitive and self-injurious behavior, temper outbursts, mood disturbances and underactivity. The physical phenotype includes distinctive facial features, hypotonia at birth, hypogonadism, small hands and feet and small stature (e.g. Cassidy & Driscoll, 2009). After an initial poor suckling relax, hyperphagia onsets in infants between the age of 1 and 6 years (Cassidy, 1997).

It is this hyperphagia that has been the focus of many studies using various methodologies, including informant based questionnaires or interviews, behavioral observations, choice paradigms and PET scanning, all of which will be reviewed briefly below. Together, the results have allowed researchers to begin to understand this internal experience in relation to specific environmental, cognitive and neurological processes and begin to identify gene-brain-behavior pathways.

Informant based interviews and questionnaires have been designed in order to gain a quantitative measure of hyperphagia using frequency or intensity of behaviors, including food 'sneaking' and theft, eating unpalatable items or taking food out of the bin. Dykens, Maxwell, Pantino, Kossler, and Roof's (2007) thirteen item informant based Hyperphagia Questionnaire considers both behavioral and emotional aspects of hyperphagia, using a scaled response for each answer to allow for comparisons both within and between individuals over time. Russell and Oliver (2003) devised the sixteen item Food Related Problem

Questionnaire that focussed upon preoccupation with food, impairment of satiety and other food-related "challenging" behavior. With robust psychometric properties (including test-retest and inter-rater reliability), Russell and Oliver recommend the use of this questionnaire for both research and clinical practice to appraise the food-related problems seen within this population. Although most individuals with Prader-Willi have the language skills to self-report some of their experiences (see Roof *et al.*, 2000), very few studies have used such reports due to issues with reliability and validity (Dykens *et al*., 2007).

Behavioral observations and choice paradigms allow for the identification of the prevalence and topography of behaviors, often in relation to groups matched on age or ability. Holland, Treasure, Coskeran, and Dallow (1995) gave individuals with Prader-Willi syndrome access to unlimited amounts of food in a laboratory setting and compared their intake with that of individuals without Prader-Willi syndrome or an intellectual disability. Individuals with Prader-Willi syndrome consumed three times more calories and took longer to show behavioral markers of satiety. The internal drive for food is so strong in individuals with Prader-Willi syndrome that they are more likely to choose a large amount of food over a small amount, even if the large amount is less-preferred (Glover, Maltzman, & Williams, 1996) or delayed (Joseph, Egli, Koppekin, & Thompson, 2002) and will even eat contaminated foods or odd food combinations (Dykens, 2000).

Recent neuroimaging work has been used to begin to understand how dysfunction in one or more areas of the brain may lead to specific behaviors. PET scanning studies have identified significant delays in activation (Shapira *et al.*, 2005) or even non-activation (e.g. Hinton *et al.*, 2006) in areas associated with satiety in typically developing controls. When asked to look at pictures of preferred food and imagine eating them, adults with Prader-Willi syndrome did not show any activation in the brain regions associated with food reward or

satiety seen in the typically developing control group. Further work using event-related-potentials suggests a difference in food stimulus evaluations between the two main genetic subtypes; those with uniparental disomys focussed more on the suitability of the food for eating, whilst those with paternal deletions focussed exclusively on the quantity of food items (Key & Dykens, 2008). This highlights the need to investigate syndrome specific behaviors in relation to genetic subtypes.

Together, the information gathered from all of these methods can be integrated to suggest a link between the underlying genetics of Prader-Willi syndrome, dysfunctional neural satiety systems and the abnormal eating behaviors (Hinton *et al.*, 2006). Such pathways are imperative to begin to understand behaviors within specific populations and consequently, implement effective interventions. In order to advance the literature around emotions and internal states within individuals with intellectual disabilities, such integration is critical.

3.4.2 Assessment of social anxiety in Fragile-X syndrome. Extremes of specific emotions or internal states are reported with several genetic syndromes associated with mild or moderate intellectual disabilities. Mood disorders have been noted within Smith-Magenis syndrome (Bersani, Russo, Limpido, & Marconi, 2007), depression within Down syndrome (e.g. Collacott, Cooper, & McGrother, 1992) and social anxiety in Fragile-X syndrome (e.g. Turk & Cornish, 1998) and Cornelia de Lange syndrome (Richards, Moss, O'Farrell, Kaur, & Oliver, in press).

Fragile-X syndrome is the most common inherited cause of intellectual disability with a current prevalence of one in 4000-6000 live births (Turner, Webb, Wake, & Robinson, 1996). There are differences in the phenotype between males and females because of the association with the X chromosome (Loesch, Huggins, & Hagerman, 2004), with males

showing a stronger phenotype and a greater degree of intellectual disability than females (Alanay *et al.*, 2007). The behavioral phenotype includes language impairment, social anxiety, gaze aversion, repetitive and self-injurious behavior (see Hagermann & Hagermann, 2002).

Again, multiple methods have been used to investigate the internal experience of anxiety within Fragile-X syndrome. Behavioral and observational methodologies have allowed researchers to conclude that individuals with Fragile-X are typically interested in social interactions, but behaviors including aversion to eye contact, self-injury, and avoidance of novel objects or situations (Turk & Cornish, 1998; Turk & Graham, 1997) highlight the difficulties they have within this area. These discomfort or avoidance behaviors can become severe enough to interfere with social functioning and peer relationships, particularly amongst females with the full mutation. Eye tracking methods have identified that individuals with Fragile-X spent less time looking at eyes and faces as a whole. However, this did not affect their ability to encode information relating to the faces (Holsen, Dalton, Johnstone, & Davidson, 2008).

Aspects of social anxiety within this population have also been investigated in terms of physiology. Galvanic skin responses increase during direct eye contact and decrease during gaze aversion (Belser & Sudhalter, 1995) and levels of cortisol are heightened following socially challenging tasks (Wisbeck *et al.*, 2000). After accounting for a number of other factors, Hessl *et al.* (2002) identified an association between salivary cortisol concentration and a number of challenging behaviors seen within children with Fragile-X, including withdrawn behavior, social problems and attention deficits (based upon carer report).

The literature is now beginning to map how the genetically derived dysfunction of several specialised neural networks (e.g. Hessl *et al.*, 2002) lead to difficulties in the ability to regulate arousal (e.g. Wisbeck *et al.*, 2000). This in turn may lead to greater reactivity to environmental and social stimuli (Miller *et al.*, 1999), making social interaction uncomfortable and promoting avoidance or escape behaviors (see Cohen, 1995, for a full discussion). Once again, this brief review has demonstrated how integrating the results of multiple methodologies can enhance our understanding and allow us to begin to infer and explore gene-brain-environment interactions of internal experiences within specific genetic syndromes.

However, we cannot rely solely upon behavioral or physiological markers to identify an internal experience. Williams syndrome is a genetically determined intellectual disability associated with a deletion on chromosome 7q11.23. It has a current prevalence of one in 20,000 and most have moderate to mild intellectual disabilities. In contrast to individuals with Fragile-X, individuals with Williams syndrome are eager to engage in social interaction, and will often do so impulsively, even with strangers (Dykens, 2003). Such behaviors persist into late adulthood (Jones *et al.*, 2000) and have been associated with specific genetically controlled neural circuitry (Meyer-Lindenberg *et al.*, 2005).

Although individuals with Williams syndrome have a high propensity to seek social interaction, like individuals with Fragile-X syndrome, they show atypical gaze behavior (albeit different in nature; Riby & Hancock, 2009). Using eye-tracking methodology individuals with Williams syndrome fixed their gaze on the actor's face for a significantly longer period than the mental age matched control group. This result was maintained for moving cartoons, but not for cartoon stills. Abnormal gaze behaviors may therefore reflect discomfort (as in Fragile-X) or intense interest (as in Williams syndrome). This dissociation

highlights the need for careful methodology and definitions alongside consideration of aetiology when observing or investigating behavior within individuals with intellectual disabilities.

3.4.3 Laughing and smiling behavior in Angelman syndrome. Angelman syndrome is a rare neurodevelopmental disorder first described in 1965 (Angelman, 1965) with a current prevalence estimate of 1 in 40,000 live births (Buckley, Dinno, & Weber, 1998; Clayton-Smith, 1993). It is caused by a disruption of the maternally inherited portion of chromosome 15q 11-13 (Clayton-Smith & Laan, 2003; Knoll, Nicholls, & Lalande, 1989) via four known genetic mechanisms (Jiang, Lev-Lehman, Bressler, Tsai, & Beadet, 1999; Lossie *et al.*, 2001). The clinical presentation of Angelman syndrome has been described in detail within the literature. The physical phenotype includes movement or balance disorder (resulting in an ataxic gait), microcephaly, epilepsy and abnormal EEG, hypopigmentation, wide mouth with widely spaced teeth, protruding tongue and high levels of salivation (resulting in drooling).

The behavioral phenotype is reviewed extensively by Horsler and Oliver (2006a). Of note are the presence of severe to profound intellectual disability, raised levels of laughing, smiling and happy demeanour, excessive sociability, little or no speech, hyperactivity and aggression in 6-10% (Summers, Allison, Lynch, & Sandler, 1995). Sixty (94%) out of the 64 studies reviewed by Horsler and Oliver identified elevated levels of laughing and smiling behaviors. The question therefore is why this behavior is present, how is it associated with the genetic cause of Angelman syndrome and how can we assess it?

Most genes have the same effect regardless of whether they were inherited from the mother or father. However, parent origin does matter for a small group of genes; a concept referred to as genomic imprinting (Murphy & Jirtle, 2003). The kinship theory (also known

as the genomic conflict hypothesis) of genomic imprinting proposes that maternal and paternal alleles can be expressed differently within the offspring in order to favour perpetuation of the maternal or paternal chromosomes (Haig & Wharton, 2003). Paternal alleles ensure survival of the offspring by increasing the cost to the offspring's mother whilst the maternal alleles promote maternal survival by reducing these costs. Maternal genes on chromosome 15q 11-13 are not fully expressed in individuals with Angelman syndrome; kinship theory therefore predicts high rates of behaviors that promote survival of the paternal chromosomes.

It has been hypothesised that some aspects of Angelman syndrome have the aim of increasing probability of gaining maternal resources (e.g. Brown & Consedine, 2004; Haig & Wharton, 2003). Using emotion-signalling theory (which suggests that expressions of emotions communicate internal states of the sender and influence the behavior of the receiver), Brown and Consedine suggest that smiling and laughing behaviors evoke high levels of social contact and, therefore, maternal resources.

It is possible to identify a number of other evolutionary benefits. Positive expressions of internal states are the primary means for social bonding between both children (Malatesta, Culver, Tesman, & Shepard, 1989) and adults (Bachorowski & Owren, 2001). As discussed in section 1.2, typically developing children who express high levels of positive internal states (such as laughing and smiling) are rated as 'cuter' (Power *et al.*, 1982) and more physically attractive (e.g. Hildebrandt, 1983) and therefore may be treated more favourably (Adams, 1977; Hildebrandt, 1982).

The focus on, and hypotheses about, the function of the laughing and smiling behaviors in children with Angelman syndrome has recently expanded. Early work suggested that these behaviors were neurologically driven, and therefore environmental

factors were not influential (e.g. Dooley, Berg, Pakula, & MacGregor, 1981; Williams & Frias, 1982). However, careful experimental manipulation of the environment identified that both the frequency and duration of these behaviors are related to environmental context.

Taking methodologies and procedures from the behavioral literature, Oliver, Demetriades and Hall (2002) conducted naturalistic observations of three children with Angelman syndrome and found that levels of laughing and smiling are significantly higher during social interaction conditions than in control conditions. Horsler and Oliver (2006b) extended this work, using analogue conditions to experimentally manipulate the environment (notably adult behavior) to observe the effect upon the child's behavior. Through this they identified that higher levels of laughing and smiling behaviors occurred when adult attention, and particularly adult eye contact, was present. Finally, employing sequential lag analyses to naturalistic observation data, Oliver *et al.* (2007) found that every smile by children with Angelman syndrome (n=13) evoked higher levels of eye contact, smiles and attention from the adult than smiles of children without Angelman syndrome (matched for degree of intellectual disability). This social contact then makes the child smile; possibly as a result of the positive affect and internal reward mechanisms initiated by these interactions (Oliver *et al.*, 2007).

Adams *et al.* (in press) extended the work of Horsler & Oliver (2006b), investigating the effect of age on smiling and laughing, looking and approach behaviours demonstrated by children with Angelman syndrome. Interestingly, the only child behaviour that changed in frequency with age was laughing and smiling which was significantly lower in the older age group than the younger age group, but only in one environmental condition; where the adult talked, touched and made eye contact. This decline in laughing and smiling behaviours with age is in line with that predicted by the genomic conflict and kinship theory (Haig &

Wharton, 2003; Brown and Consedine, 2004). Typically as a child ages and requires less maternal resources, it might be hypothesised that these resource-soliciting behaviours will decrease. It could be hypothesised that as the child with Angelman ages, they find social interaction and eye contact less reinforcing, it therefore evoke less positive affect and subsequently less laughing and smiling behaviours in older children.

Therefore, the smiling and laughing behaviors seen in Angelman syndrome are a highly efficient way of promoting survival of the paternal chromosomes as they are both beneficial to the child (increases investment from mother) and costly to the mother (time lost pursuing the welfare of other children or activities). Robust methods of assessing these behaviors are now being used and, consequently, are providing more information on the exact topography and potential function in relation to genetics and, specifically, genomic imprinting. Further research and integration of genetics, neuroimaging, physiology and behavioral observations may allow us to further understand the gene-brain-behavior pathway. However, due to the inability to self-report, the question remains as to whether these laughing and smiling behaviors reflect an internal experience of positive affect.

3.4.4 Summary

By identifying and reviewing the literature pertaining to the assessment of internal states (hunger, social anxiety, laughing and smiling) within specific genetic syndromes, it has been possible to identify multi-level approaches to assessing associated behaviours. Some of these, such as scanning techniques and eye movement data, are arguably more likely to be used in research than clinical settings. Nonetheless, they have provided essential information for understanding these behaviours from both a neurological and physiological perspective. Other studies, such as those that have identified the distinct pattern of behavior demonstrated by individuals with Fragile-X upon meeting strangers (which highlight the internal

experience of social anxiety), questionnaires which focus upon hunger related behaviour in Prader-Willi syndrome, and those that identified the association between social contact and higher levels of smiling and laughing behaviours in Angelman syndrome, used methods that have excellent potential to be used clinically. A common thread of all of the studies reviewed within this section is that they use carefully designed measures with excellent reliability and validity which focus upon a narrow range of internal experience rather than attempting to assess a broad range of behaviours relating to multiple internal experiences.

4 Conclusions

This review has demonstrated the importance of observing and understanding behavioral and facial expressions of internal states in individuals with severe or profound intellectual disabilities. These expressions provide valuable insights into the internal experiences of individuals who cannot directly communicate their thoughts and feelings. The paucity of literature pertaining to the emotions of individuals with severe or profound intellectual disabilities is of concern. Only by expanding the remit of this review to encompass broader internal states could methods of assessment be identified and evaluated.

Examination of the literature regarding the expression of internal states in individuals with severe or profound intellectual disabilities illustrated several points. Firstly, that research within this area is limited. Secondly, that there may be deficits in both the sending and receiving of facial expressions of internal states within this population. Individuals with severe or profound intellectual disabilities may exhibit more subtle facial expressions of internal states, which adults are poor at interpreting if they do not have experience of caring for, or communicating with, this population. Such difficulties were also noted within the extremes of internal states, notably depression, anxiety and pain. Although symptoms may be slightly atypical it is important to remember that they may reflect the same level of

internal experience (and arguably discomfort) and therefore should be responded to appropriately.

By reviewing the syndrome specific literature, it has become apparent that there are numerous measures and methods that have been designed or adapted to investigate and explore various internal states among individuals with intellectual disabilities. All of the approaches used, including informant based measures, observational or experimental designs, choice paradigms, event-related potential work and neuroimaging studies have demonstrated potential methods of assessment with robust reliability and validity and the understanding of gene-brain-behavior pathways for each specific syndrome.

In relation to emotion and internal states, investigating syndrome specific differences suggests support for underlying genetic mechanisms which determine these specific pathways. This highlights the importance of considering the aetiology of an individual's intellectual disability when considering internal and emotional states and experiences. For example, whilst eye contact may be highly potent and evoke laughing and smiling in individuals with Angelman syndrome, this finding cannot be generalised to the wider population of individuals with intellectual disabilities as some individuals may find it less reinforcing or even uncomfortable (e.g. those with autism or Fragile-X (e.g. Knivsberg, Reichelt, Høien, & Nødland, 2002)). There is no reason to suggest that this interplay between genetics and expression of internal states should be unique to the syndromes discussed within this review, and indeed, there is emerging literature of increased internal states within other genetic syndromes associated with severe or profound intellectual disabilities (e.g. Cornelia de Lange syndrome; Richards *et al.*, in press).

Overall, it is apparent that the knowledge and literature base is at its peak in areas where there has been integration within and across methodologies and subject areas such as

anxiety, depression, pain and the syndrome-specific literature. It therefore is imperative that research within the area of emotional expression in individuals with severe or profound intellectual disabilities strives not only to enhance knowledge but also to inform and inspire the use of multiple approaches. We will not be successful at understanding and intervening in complex problems or behaviors until we begin to generate multilevel models (Oliver & Woodcock, 2008). Only when this has been established can behavior begin to be understood in terms of its association with the environment, the brain and ultimately, the individual's genetics. This level of understanding can then promote person-centred, empirically supported interventions that recognise the importance of emotional and internal states at an individual level.

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